Scabies masquerading as Letterer-Siwe's disease

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Letterer-Siwe's disease was diagnosed from clinical appearance and initial assessment of a skin biopsy in a child with a 2-month history of skin rash. Fine erythematous papules were scattered on the trunk. The biopsy showed epidermal thickening and an inflammatory infiltrate chiefly in the upper layers of the dermis; deeper in the dermis the infiltrate was perivascular and periappendicular, histiocytes predominating in some areas and lymphocytes in others. A diagnosis of scabies was made after burrows were demonstrated on palms and soles and the mite of scabies was isolated from them.

Un diagnostic de maladie de Letterer-Siwe a été posé à partir du tableau clinique et de l'évaluation préliminaire d'une biopsie cutanée chez un enfant présentant une éruption cutanée d'une durée de 2 mois. De fines papules érythémateuses étaient disséminées sur le tronc. La biopsie a révélé un épaississement de l'épiderme et une infiltration inflammatoire, principalement dans les couches supérieures du derme; plus profondément dans le derme des infiltrats périvasculaires et périappendiculaires ont été observés, avec prédominance des histiocytes dans certaines régions et des lymphocytes dans d'autres. Un diagnostic de gale a été posé après avoir démontré la présence de fistules sur les paumes et les plantes, desquelles on a pu isoler l'acare de la gale.

The skin rash of Letterer-Siwe's disease, with its "prodigious . . . variety of appearance",[1] has frequently been mistaken for other cutaneous disorders. Conversely, skin conditions in children are not infrequently diagnosed incorrectly as "Letterer-Siwe's disease". Scabies is unlikely to be a source of confusion, although dermatologists have been aware that scabies can compete with syphilis as a mimic of other diseases. However, one case of suspected Letterer-Siwe's disease, reported below, was shown to be scabies, and expensive and unnecessary investigation, treatment and parental anxiety were thereby avoided.

Case report

A 13-month-old boy was admitted to Izaak Walton Killam Hospital for Children with a widespread rash that had developed 2 to 3 months previously. Just prior to the onset of the eruption the child had been admitted to a hospital in his home area because of otitis media. The eruption had been treated at home with an unknown medication but, because it did not respond to this treatment, the child was seen by a pediatrician, who advised a skin biopsy. From the biopsy findings the child was suspected to have Letterer-Siwe's disease. He was therefore referred to our hospital.

The child looked healthy, played happily and ate well. His temperature was normal. Other than slight enlargement of some lymph nodes no physical abnormalities were found. Neither the liver nor the spleen was enlarged. No abnormal hematologic findings were reported. However, the skin rash prompted a dermatologic consultation, particularly since the skin biopsy examined previously had been reported as suggesting Letterer-Siwe's disease. Scattered fine, erythematous papules, some larger and orange, were noted on the trunk. There was no purpura. The scalp was normal, showing neither the copper papules of Letterer-Siwe's disease nor seborrhea. The palms and soles, however, contained at least three burrows (Fig. 1), from which the mite of scabies was isolated.

In one of two biopsy slides submitted from the referring hospital focal thickening of the epidermis was seen, accompanied by an infiltrate chiefly in the upper layers of the dermis (Fig. 2). In some areas the infiltrate encroached on the dermoepidermal junction and invaded the epidermis (Fig. 3). In addition there was a perivascular and periappendicular infiltrate in the deeper layers of the dermis, extending into the subcutaneous fatty layers. This infiltrate, which at first appeared pleomorphic, consisted of a large but variable number of lymphocytes, eosi-

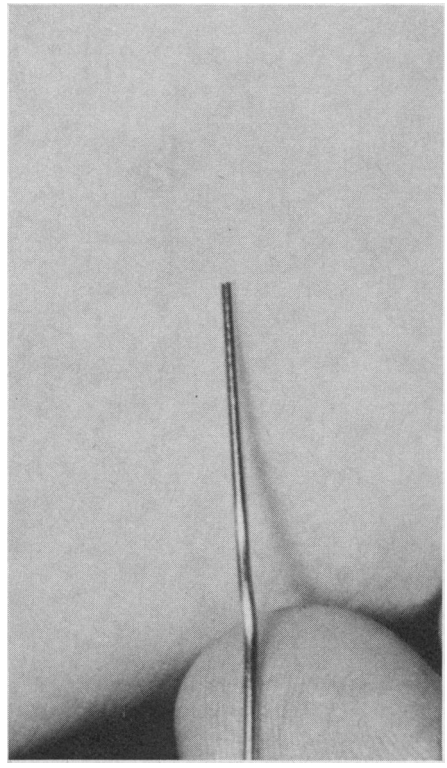

FIG. 1—Mite burrow on sole of child with scabies.

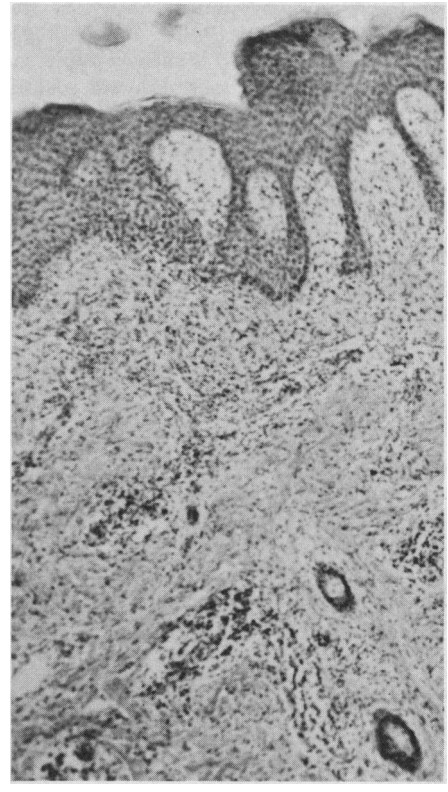

FIG. 2—Survey of skin to show inflammatory infiltrate mainly in upper dermis, with perivascular and periappendicular inflammatory cell aggregates in deeper parts (hematoxylin—eosin [H—E]; x40).

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nophils and histiocytes. While in some areas histiocytes predominated, in others, particularly the periappendicular regions, lymphocytes predominated (Fig. 4). Only a few mitoses were seen.

While there were some disturbing features that, presumably, had led to the diagnosis of Letterer-Siwe's disease, other features in one of the biopsy sections were not compatible with such a diagnosis — namely, the epidermal changes, the predominance of lymphocytes and the pattern of distribution of the inflammatory cells. Fortunately, the demonstration of the mites on clinical examination confirmed the diagnosis of an inflammatory (hypersensitivity) response in a case of scabies.

By this time a second skeletal radiographic survey had been carried out; no abnormalities were detected. The first skeletal survey, in which 14 radiographs were taken, had been performed before referral. Following treatment with gamma banzine hexachloride (Kwellada) the child was discharged after arrangements had been made to treat all persons in his household.

Discussion

Winkelmann,[2] reviewing the skin changes in histiocytosis X (the term embracing the three conditions Letterer-Siwe's disease, Hand-Schüller-Christian disease and eosinophilic granuloma), noted that extensive skin lesions are the rule in Letterer-Siwe's disease. The changes in the skin produced by the epidermal hyperplasia and histiocytic infiltration vary depending on the amount, rapidity and intensity of the reaction. Necrosis with hemorrhage, erosion and crusting is seen in rapid infiltration. Papules, nodules and scaling are seen in slow infiltration. Clinical appearances have been classified by Altman and Winkelmann[3] and include a diffuse papular or scaly "seborrheic" eruption, a petechial purpuric eruption, granulomatous ulcerative lesions, xanthomatous lesions and bronzing of the skin.

Seborrheic dermatitis, Darier's disease (keratosis follicularis) and refractory eczema must be excluded in the differential diagnosis,[2,4] but scabies is not usually considered. The possibility of misdiagnosing the lesions of nodular scabies as those of a form of cutaneous lymphoma or reticulosis on histologic grounds alone was stressed by Thomson and colleagues.[5] Although the presence of burrows with mites is the primary diagnostic sign in scabies, the secondary eruption can predominate and cause confusion. Possibly as a result of sensitization, urticated papules can develop about the trunk and may become excoriated and crusted. Excoriation can cause cutaneous hemorrhage that appears similar to the bronze purpura of Letterer-Siwe's disease. However, the patient with scabies usually looks well; this feature, as in our patient, should weigh heavily against the diagnosis of Letterer-Siwe's disease.

Clinical examination of the infant's skin should always include palms, soles and genitalia. As opposed to adults and older children, children up to approximately 3 years of age will commonly have burrows with mites in these areas.[6,7] Occasionally on the palms and soles the lesions are bullous and are confused with the vesicular dermatitis of infantile eczema. In common with adults, erythematous papules are often noted on the shaft of the penis and on the scrotum in infants. The occasional persistence for several months of itching erythematous nodules and papules may also be confusing.[8,9] Although the histologic picture in scabies has some features in common with that in Letterer-Siwe's disease, the intense inflammatory reaction in the epidermis, with a preponderance of lymphocytes but only a few larger mononuclear and reticular cells, should help in the distinction of these lesions. The few papules are widely distributed and the intervening skin is normal in this form of scabies. Reticulosis may also be mimicked when topical corticosteroids have been applied unwittingly to a patient with scabies.[10]

Demonstration of the mite, to establish the diagnosis of scabies, is relatively easy, since the mite can be lifted out from the end of the burrow on a sewing needle. Other simple methods include placing a drop of 10% potassium hydroxide in water, saline or mineral oil over a suspected burrow, which is then gently scraped.[11,12] The debris should contain the mite and eggs. While a biopsy can occasionally demonstrate the mite in the skin,[13] this procedure is not necessary.

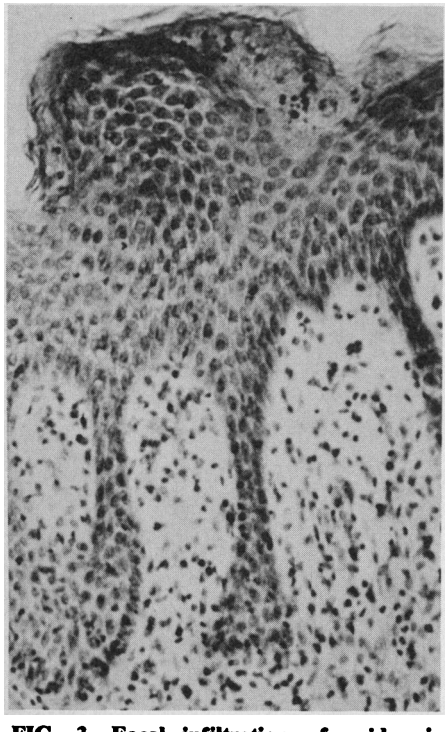

FIG. 3—Focal infiltration of epidermis and diffuse infiltration of dermis by inflammatory cells (H–E; x100).

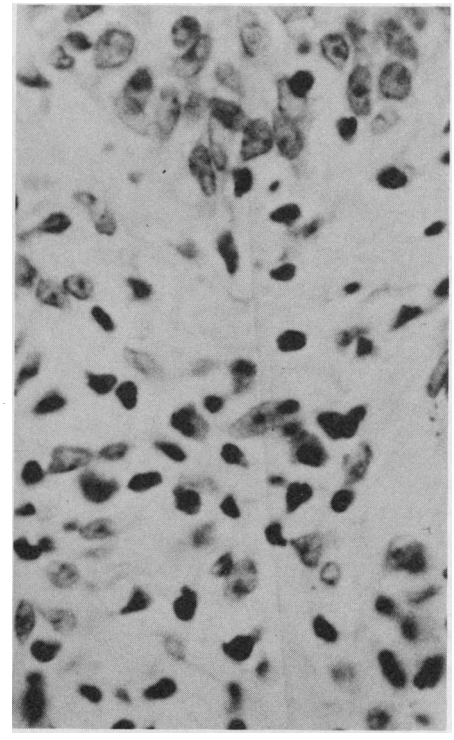

FIG. 4—Pleomorphic infiltrate near dermoepidermal junction, consisting chiefly of lymphocytes, histiocytes and neutrophils. An occasional mitosis was seen (H–E; x600).

Conclusion

Although histologic features can be confusing, correlation of the finer clinical and pathological details, best achieved by joint consultation of pathologist and dermatologist, should allow differentiation of scabies and Letterer-Siwe's disease.

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